Eclamptic seizures causing fractures appear to be very rare. However, any post eclamptic patient who complains of hip pain should be thoroughly assessed, including the use of X-rays, expert radiologist review and orthopaedic assessment. Distracting pains, such as the severe headache experienced by our patient, may lead to a delay in the diagnosis. Subcapital femoral fractures ought to be considered in the absence of more common fractures or other obvious abnormalities. If in doubt, any abnormal hip X-rays should be compared with the normal side to exclude the presence of a fracture. Hip pain as such is not a complication of Caesarean section or vaginal delivery and the cause should be sought.

Transient osteoporosis of the hip is an uncommon condition and has two demographic peaks, one during the third trimester of pregnancy and the other between the fifth and sixth decade of life. This painful regional osteoporosis affects previously healthy women in the third trimester of pregnancy. It is characterized by pain in the affected hip and pronounced osteopenia of the femoral head and neck. Radiographs show pronounced osteopenia of the femoral head and neck with preservation of the joint space. Bone scan and magnetic resonance imaging are sensitive but not specific for diagnosis, and laboratory studies are typically normal. It has a relatively short clinical course (average 6 months) and a predictably benign prognosis. Complete clinical and radiological recovery is the rule. The diagnosis is one of exclusion. The cause of the osteopenia is not known, although various aetiological factors have been implicated.

In summary, aggressive management to minimize the risks of seizures in pre-eclamptics are essential in preventing the complications we have reported here. Clinicians need to be aware of the potential for hip fractures, including bilateral subcapital fractures, during eclamptic seizures.

References

Rocuronium for muscle relaxation in two children with Friedreich’s ataxia

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Friedreich’s ataxia is a rare hereditary neurodegenerative disease caused by a defect in the gene that encodes a mitochondrial protein called frataxin. We report the use of rocuronium 0.6 mg kg⁻¹ in two adolescent girls with Friedreich’s ataxia undergoing propofol–sufentanil–oxygen–air anaesthesia for spinal surgery. Neuromuscular transmission was monitored using acceleromyography, and onset and recovery times were recorded. The clinical duration of rocuronium was comparable to that of children without neuromuscular disease (25% recovery T₁=44 and 24 min for patients 1 and 2 respectively).

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Friedreich’s ataxia is a rare hereditary, autosomal recessive disease that causes progressive degeneration of the spinocerebellar and pyramidal tracts. The main complaint is progressive limb and gait ataxia with skeletal muscle weakness. Patients with Friedreich’s ataxia are managed during anaesthesia in the same way as patients suffering from amyotrophic lateral sclerosis. The latter is also a neurodegenerative disease in which there is marked sensitivity to non-depolarizing neuromuscular blocking drugs (NDNMB) and a tendency to hyperkalaemia after administration of succinylcholine. Information about the anaesthetic management of patients with Friedreich’s ataxia is anecdotal. Reports on the response to NDNMB are conflicting and vary from marked sensitivity to a normal activity to non-depolarizing neuromuscular blocking drugs (NDNMB) and a tendency to hyperkalaemia after administration of succinylcholine. Most notably, in nearly all reports anaesthesia was supplemented with volatile anaesthetics, which are known to produce dose-dependent increases in the magnitude and duration of neuromuscular block. We describe two patients with Friedreich’s ataxia who underwent posterior spinal fusion under total i.v. anaesthesia where rocuronium was used successfully for muscle relaxation.

### Case reports

**Patient 1**

A 15-yr-old girl, height 167 cm and weight 71 kg, presented for general anaesthesia for scoliosis repair. Friedreich’s ataxia was diagnosed clinically at the age of 7 yr, and 2 yr later was confirmed by genetic testing. She was confined to a wheelchair by the age of 13 yr because of muscle weakness and progressive thoracolumbar kyphoscoliosis. She had a history of recurrent tachydysrhythmias and took verapamil orally. She had no history of previous anaesthesia or surgery.

Physical examination revealed marked kyphoscoliosis. She presented as a normally developed girl with slightly reduced muscle strength. The electrocardiogram showed sinus rhythm, and QRS and T patterns consistent with left ventricular strain. Echocardiography revealed concentric left ventricular hypertrophy without obstruction and with good myocardial function. The pulmonary function tests showed an early restrictive abnormality, the forced vital capacity being 2.2 litres (predicted value 3.6 litres) and the forced expiratory volume in 1 s 2.1 litres (predicted 3.0 litres). Transcutaneous oxygen saturation was 95% with ambient air.

After written informed consent from the parents, the girl was fasted overnight and premedicated orally with dikalium-chlorazepat (a benzodiazepine) 10 mg. After the placement of i.v. access and routine monitors (pulse oximeter, ECG, non-invasive arterial pressure monitoring), pre-oxygenation and administration of glycopyrrolate 0.004 mg kg⁻¹ i.v., anaesthesia was induced with propofol 2.5 mg kg⁻¹ and sufentanil 0.5 µg kg⁻¹. After adequate mask ventilation had been established, the trachea was intubated without muscle relaxation and the lungs were ventilated with oxygen in air. A double-lumen catheter was inserted into the right internal jugular vein and the left radial artery was cannulated. A urinary catheter attached to an hourly measuring chamber was inserted. Anaesthesia was maintained with an i.v. infusion of propofol 100–200 µg kg⁻¹ min⁻¹ and sufentanil 0.01–0.03 µg kg⁻¹ min⁻¹, titrated to effect. Volatile anaesthetics were not used. For perioperative antibiotic prophylaxis, cephalozolin 2 g was given i.v. Tidal volume and respiratory rates were controlled to keep the arterial carbon dioxide pressure between 35 and 40 mm Hg. A warm forced air device was used to maintain body temperature at 36°C, which was monitored by a bladder probe. A device for intraoperative cell saver blood salvage was set up.

After placing the patient in the prone position on the foam-padded operating table, the patient’s right arm was prepared for neuromuscular monitoring using acceleromyography (TOF-Watch SX; Organon, Germany). The ulnar nerve was stimulated supramaximally with repeated train-of-four (TOF) stimuli (2 Hz, 0.2 ms duration at 15 s intervals) using surface electrodes placed above the wrist. The transducer was tightly fixed to the distal interphalangeal joint of the thumb. An i.v. infusion and arterial pressure cuff were not inserted in the monitored arm. The TOF monitor was connected to a portable PC for online data recording and processing (TOF-Watch SX Monitor program; Organon, Dublin, Ireland). After calibration and initial signal stabilization of the control response, rocuronium 0.6 mg kg⁻¹ was given i.v. over 5 s. After rocuronium administration, the heart rate increased from 75 to 80 beats min⁻¹. The time from administration of rocuronium to complete clinical recovery was monitored. In accordance with the Copenhagen Consensus Conference, the following time course of neuromuscular block was measured: (i) onset time; (ii) time of recovery of first twitch of the TOF response to 10, 25 and 90%; (iii) recovery index (time between 25 and 75% recovery of first twitch of TOF); and (iii) recovery time (time between 25% recovery of first twitch and recovery of TOF ratio to 90%). On the request of the surgeons, an incremental dose of rocuronium 0.2 mg kg⁻¹ was given 75 min later as T₁ had returned to...
100% and the TOF to 90%. The response to TOF stimuli after the increment was also recorded. The clinical data are summarized in Table 1. Figure 1 shows the recording of the TOF response to the initial and incremental dose of rocuronium in this patient.

The surgical procedure lasted 6.5 h. The estimated blood loss was 1 litre and this was replaced with one pack of allogenic red blood cells and 250 ml autologous cell-saver salvaged blood and 3 units of fresh frozen plasma. The arterial pressure remained stable throughout the procedure. The girl was artificially ventilated overnight. On the morning of the following day, the tracheal tube was removed without problems. The remainder of her postoperative course was uneventful.

**Table 1** Time course of neuromuscular block after administration of rocuronium 0.6 mg in the two patients. TOF<sub>90</sub>=ratio of fourth to first twitch of train-of-four (TOF) response; T<sub>10</sub>, T<sub>25</sub>, T<sub>90</sub>=time to recovery of the first twitch of TOF to 10, 25 and 90 min respectively; T<sub>25</sub>±T<sub>75</sub>=time for 25% to 75% recovery of first twitch of TOF; T<sub>25</sub>±TOF<sub>90</sub>=time for 25% recovery of the first twitch to a TOF ratio of 90%.

<table>
<thead>
<tr>
<th>Patient</th>
<th>Onset time (min)</th>
<th>T&lt;sub&gt;10&lt;/sub&gt; duration (min)</th>
<th>T&lt;sub&gt;25&lt;/sub&gt; duration (min)</th>
<th>T&lt;sub&gt;90&lt;/sub&gt; duration (min)</th>
<th>T&lt;sub&gt;25&lt;/sub&gt;±T&lt;sub&gt;75&lt;/sub&gt; recovery index (min)</th>
<th>T&lt;sub&gt;25&lt;/sub&gt;±TOF&lt;sub&gt;90&lt;/sub&gt; recovery time (min)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Patient 1</td>
<td>1.25</td>
<td>38.75</td>
<td>44.5</td>
<td>62.5</td>
<td>12</td>
<td>27.25</td>
</tr>
<tr>
<td>Incremental dose 0.2 mg</td>
<td>1.5</td>
<td>25.5</td>
<td>30.75</td>
<td>45.5</td>
<td>11</td>
<td>30.75</td>
</tr>
<tr>
<td>Patient 2</td>
<td>0.9</td>
<td>20.4</td>
<td>23.9</td>
<td>42.4</td>
<td>13.25</td>
<td>36.75</td>
</tr>
</tbody>
</table>

**Discussion**

We present, for the first time, information concerning the response to a standard dose of rocuronium in children with Friedreich’s ataxia. After administration of 0.6 rocuronium mg kg<sup>−1</sup>, we found in one patient (no. 2) onset and recovery data comparable to published data from children without neuromuscular disease, whereas in the other patient neuromuscular monitoring revealed a prolonged recovery time.

Friedreich’s ataxia is a rare, recessively inherited disease with a variety of causes for its pathogenesis. Recent advances in basic research have revealed a mutation in the gene that encodes the mitochondrial protein frataxin as the underlying pathological cause. Reduction in frataxin protein production leads to mitochondrial iron accumulation with impaired mitochondrial respiratory function.

The main anaesthetic implications of Friedreich’s ataxia relate to concomitant myocardial problems and the administration of neuromuscular blocking drugs. Early reports of hyperkalaemia after administration of succinylcholine to patients with disorders resulting in denervated muscle suggest that it is preferable to avoid depolarizing muscle relaxants in these patients. There are only a few reports of the use of non-depolarizing muscle relaxants in patients with Friedreich’s ataxia. Though an early case report showed hypersensitivity to tubocurarine, later anecdotal reports described a normal or near-normal response to various non-depolarizing neuromuscular blocking drugs (tubocurarine, atracurium, vecuronium). In all the case reports, anaesthesia was supplemented with volatile anaesthetics (isoflurane, enflurane or halothane), agents known...
for their enhancing effect on neuromuscular block, which can result in prolonged recovery. Only one published report gives complete details of neuromuscular monitoring after administration of atracurium.5

A bolus of rocuronium 0.6 mg kg\(^{-1}\) has an onset time of approximately 90 s and an intermediate clinical duration (T\(_{25}\)) of about 25 min, with a wide individual range. Though the clinical duration (T\(_{25}\)) differed markedly between our two patients (44 and 24 min for patients 1 and 2, respectively; Table 1), the data are within the range reported in children without neuromuscular disease.8 9 The time course of neuromuscular block and recovery in the two patients confirms the report by Mouloudi and colleagues, who found normal recovery data after atracurium in a woman with Friedreich’s ataxia. However, as a result of the administration of a different chemical type of neuromuscular blocking drug by these workers and the supplementary use of isoflurane in the earlier report, comparison with our report is of limited use. The recovery indexes in patient 1 (12 min) and patient 2 (13 min) are comparable to data from patients without Friedreich’s ataxia, indicating normal pharmacokinetics in these patients.

Comparison of neuromuscular block data in patients with neuromuscular disease is difficult. In addition to a comparable study design with respect to anaesthesia and drug dose, the neuromuscular monitoring techniques, such as the method of measurement, frequency of stimulation and the site of neuromuscular monitoring, are important. All these factors may influence evoked responses to electrical stimuli. In our investigation neuromuscular monitoring was performed using acceleromyography. We chose acceleromyography because of its ease of use compared with bulky mechanomyography equipment during a procedure in which the patient was in the prone position compared with the fairly bulky equipment of mechanomyography. Another option would have been the use of electromyography. Though an investigation in children without neuromuscular disease showed the superiority of acceleromyography compared with electromyography, monitoring recovery from residual neuromuscular block in patients with Friedreich’s ataxia is not well described.10

In contrast to reported cases where volatile anaesthetics were administered, we chose an anaesthetic technique based on propofol and sufentanil titrated to effect without any inhalational agent. This anaesthetic technique, together with adequate fluid replacement, enabled us to avoid major changes in arterial pressure. Both patients had left ventricular hypertrophy without outflow obstruction, a common finding in patients with Friedreich’s ataxia.11 As both girls had normal left ventricular function, no other anaesthetic measures were required during surgery.

In summary, the successful anaesthetic management of two children with Friedreich’s ataxia scheduled for major orthopaedic surgery is reported. The response to a bolus of rocuronium 0.6 mg kg\(^{-1}\) was comparable to its effect in children without neuromuscular disease, although a greater number of patients is required to confirm these findings. We recommend accurate assessment of neuromuscular block throughout anaesthesia in patients with Friedreich’s ataxia.

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